structures were predominant: they are usually absent in middle ear adenomas. In these cells the nuclei were midcellular and apical, as reported in papillary adenocarcinomas.1,2 Recent and old haemorrhages were seen associated with areas of fibrosis and reactive changes. Iron positive granules in tumour cells and erythrocytes within gland lumina were conspicuous. This is a common feature of papillary carcinomas and serous gland tumours which rarely invade the temporal bone.6 Immunohistochemical results were consistent with those of the few previous reported studies. Tumour cells are positive with keratins, frequently positive with S-100, and less often with neuronal markers.7,8 EMA and vimentin have also been found to be positive, with vimentin staining restricted to the basal portion of cells.7

Papillary adenocarcinomas of temporal bone are characterised by local invasion with bone destruction, facial nerve involvement, and extension both to the posterior cerebellar fossa and the internal auditory canal. This regional aggressiveness is related to the histogenesis of the lesion, the origin of which has been debated. However, the derivation from endolymphatic sac epithelium proposed by Heffner9 seems to be accepted now. The endolymphatic sac originates from the neuroectoderm (otocyst). It is situated between the dura and the posterior surface of the petrous portion of the temporal bone. Its epithelium is arranged in villous folds and lies on loose connective tissue containing blood vessels. These topographical and microscopic characteristics are strong arguments for an endolymphatic sac origin for this tumour as opposed to one of the middle ear. This further observation leads us to insist on the differentiation of this tumour from adenomas or mixed tumours arising in the middle ear cleft. The other point of interest is the association with VHL disease. A few such cases have been reported.10,11 In three of them the lesion was bilateral.1,5 This argues for recommending the inclusion of papillary carcinoma of the endolymphatic sac in the spectrum of neoplasms seen in VHL disease. Moreover, this otological manifestation can be the initial sign of the disease.3

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**Endochondral pseudocyst of the auricle**

J A Lee, A Panarese

**Abstract**

Endochondral pseudocyst of the auricle is an uncommon, though distinctive clinicopathological entity occurring mainly in young men. An additional case is reported and the differential diagnosis and pathogenesis discussed. It is suggested that lymphatic dilatation of normally present tissue planes could be the most likely causative mechanism. Minor trauma to susceptible ears also seems to be a requirement for development of this condition.

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Endochondral pseudocyst of the auricle is an uncommon condition which presents as a painless, dome-shaped, cystic swelling on the anterior surface of the upper part of the auricle.1 Although originally reported in Chinese people,2 it can occur in any race.3 All ages may be affected, but the lesion occurs primarily in young adults and shows a strong male predominance (90% of cases). Ten to 20 per cent of patients develop a similar lesion on the contralateral pinna, usually asynchronously. A needle aspirate produces 0.5-10 ml of straw coloured, viscous, albumin-containing fluid with osmolarity, glucose, and protein concentrations similar to those of serum.1,3
Microscopically there is an intracartilaginous cyst which does not have an epithelial lining—hence the designation pseudocyst. Because of its relative rarity, the condition is frequently misdiagnosed by clinicians and pathologists. The pathogenesis also remains unclear.

Case report
A 32 year old man was referred to clinic with an 18 month history of a localised swelling on the left ear. It had increased slightly in size over this period and was asymptomatic apart from slight tenderness and throbbing after sleeping on that side. There was no history of ear disease or trauma and his medical history was also unremarkable. Physical examination showed a firm, subcutaneous swelling 2 cm in diameter, located between the helix and antihelix in the upper third of the left auricle. A clinical diagnosis of chondroma was made and an excision biopsy was performed. A small incision was made just above the swelling; skin and perichondrium were separated from the cartilage, and the lesion was removed. A small suction drain was left in situ for 24 hours and a pressure bandage was also applied for this period. The patient was discharged the following day. A month later, he was symptom free, the wound had healed, and there was no deformity of the auricle. The right ear looked normal.

The surgical specimen consisted of two fragments of cartilage measuring 15 × 10 × 2 mm and 5 × 4 × 2 mm; macroscopic central cyst formation was visible. Microscopic analysis showed central cystic degeneration of the auricular cartilage (fig 1). In places the cartilage had a ragged edge, but in most areas the cyst had a lining composed of amorphous tissue containing scattered cells. Although this had an appearance suggestive of fibrous connective tissue on haematoxylin and eosin staining, special stains showed no evidence of collagen or elastin fibres and immunohistochemical staining for S100 protein showed that the scattered cells were chondrocytes. Thus most of the cyst lining represents degenerating cartilage. Alcian blue staining with and without hyaluronidase showed that the degenerating cartilage had a high content of acid mucins; however, little mucin was present in the cystic spaces. Intracystic granulation tissue was absent in this case. Immunohistochemical staining for low and high molecular weight cytokeratins was negative, confirming the absence of an epithelial lining. However, some of the spaces showed a discontinuous lining which stained with the endothelial marker factor VIII (fig 2).

Discussion
Endochondral pseudocyst of the auricle is a distinctive condition presenting as a swelling of the upper part of the auricle. The clinical differential diagnosis is as wide-ranging as that of a lump in the ear, including conditions such as chondroma, fibroma, epidermoid cyst, chondrodematitis nodularis helicis, gouty tophus, haemangiomia, and even angiosarcoma. In practice diagnosis depends largely on awareness of the condition, as insertion of a needle will allow the contents to be aspirated with subsequent subsidence of the swelling. Notwithstanding earlier reports of recurrence following aspiration, this simple, relatively non-invasive approach, coupled with pressure bandages or steroid injection, can cure at least a proportion of cases. When an excision is performed the histological appearances of the surgical specimen are distinctive and again diagnosis rests largely on awareness of the condition. Lesions for which endochondral pseudocyst is most commonly mistaken include relapsing polychondritis, chondrodematitis nodularis helicis, cauliflower ear, and subperichondrial haematoma. Although differential diagnosis might be difficult in a fragmented specimen, an essential point of difference between endochondral pseudocyst and the conditions cited is that the lesion is intracartilaginous in pseudocyst, but subperichondrial in the others. Relapsing polychondritis is painful and diffuse, in contrast to the localised, painless swelling of endochondral pseudocyst. In severe chondrodematitis nodularis helicis granulation tissue can penetrate the cartilage and cause cystic dilatation, but both sides of the cartilage plate and overlying skin are involved, in contrast to endochondral pseudocyst, where the outer cartilage plate and skin are normal. Although atypical chon-
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... fluid contains solute concentrations similar to those of serum and includes albumin, findings which indicate that the spaces may develop from dilated lymphatics. Our novel observation of a discontinuous factor VIII positive lining to the pseudocyst in the present case is consistent with this interpretation. In keeping with a predisposing congenital factor is the bilateral occurrence of the condition in many cases.

If potential spaces are as common in adult ears as is suggested in the cited study (a point which would benefit from further investigation), then endochondral pseudocyst should also be extremely common. We suggest that an additional factor, such as "low grade trauma" is required as a stimulus for the spaces to open up. An observation which supports this hypothesis is the finding of auricular pseudocysts (bilateral in two cases) in four children with severe atopic eczema. All four patients had eczema on their ears and it was considered that minor trauma from rubbing and scratching may have played a part in pseudocyst development. It seems likely that repeated minor trauma to the auricle in children will open up residual tissue planes more easily than in adults, in whom they have been closed for longer.

In conclusion, endochondral pseudocyst is a distinctive clinicopathological entity which it is relatively straightforward to recognise once its existence is appreciated. We suggest that the most likely pathogenesis is re-opening of normally present tissue planes by lymphatic dilatation; an additional stimulus such as repeated minor trauma to susceptible ears also seems to be a requirement for development of the condition.